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cont  
b) those described in Table 3 (SEQ ID NO:18-SEQ ID NO:67); and

c) those including the introns-exons junctions of Table 2;

d) those derived from the primers in a), b), or c).

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**REMARKS**

Favorable consideration of this application and entry of the foregoing amendments are respectfully requested.

The specification has been amended to make reference to certain sequence identifiers and to include the Sequence Listing submitted herewith on separate sheets. Entry of the Sequence Listing does not raise the issue of new matter as the sequence information contained therein is presented in the application as originally filed. The computer readable copy of the Sequence Listing submitted herewith is believed to be the same as the attached paper copy of that Listing.

Attached hereto is a marked-up version of the changes made to the specification and claims by the current amendment. The attached page/s is/are captioned "**Version With Markings To Show Changes Made.**"

An early and favorable Action on the merits is  
requested.

Respectfully submitted,

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VERSION WITH MARKINGS TO SHOW CHANGES MADE

IN THE SPECIFICATION:

The paragraph beginning at page 6, line 1:

Figure 2: Sequence of the human nCL1 cDNA (B), and the flanking 5' (A) and 3' (C) genomic regions.

A) (SEQ ID NO:68) and C) (SEQ ID NO:69) The polyadenylation signal and putative CAAT. TATAA sites are boxed. Putative Sp1 (position -477 to -472), MEF2 binding sites (-364 to -343) and CArG box (-685 to -672) are in bold. The Alu sequence present in the 5' region is underlined.

IN THE CLAIMS:

1. (Amended) A nucleic acid sequence comprising:
  - 1) the sequence represented in Figure 8 (SEQ ID NO:1-SEQ ID NO:4); or
  - 2) the sequence represented in Figure 2 (SEQ ID NO:5, SEQ ID NO:68 and SEQ ID NO:69); or
  - 3) a part of the sequence of Figure 2 (SEQ ID NO:5, SEQ ID NO:68 and SEQ ID NO:69) with the proviso that it is able to code for a protein having a calcium dependant protease activity involved in a LGMD2 disease; or

4) a sequence derived from a sequence defined in 1), 2) or 3) by substitution, deletion or addition of one or more nucleotides with the proviso that said sequence still codes for said protease.

4. (Amended) A nucleic acid sequence encoding the aminoacid sequence represented in Figure 2 (SEQ ID NO:6).

6. (Twice Amended) An aminoacid sequence according to claim 5 characterized in that either it contains the sequence such as represented in Figure 2 (SEQ ID NO:6), or the amino acid sequence of Figure 2 (SEQ ID NO:6) modified by deletion, insertion and/or replacement of one or more amino acids with the proviso that such aminoacid sequence has the calpain activity involved in LGMD2 disease.

15. (Amended) A method for detecting of a predisposition to a LGMD2 disease in a family or a human being, such method comprising the steps of:

- selecting one or more exons or their flanking sequences of the gene,
- selecting primers specific for these exons, or their flanking sequences, or an hybrid thereof,
- amplifying the nucleic acid sequences with these

primers, the substrate for this amplification being the DNA of a human being; and

- comparing the amplified sequence to the corresponding sequence derived from Figure 2 (SEQ ID NO:5, SEQ ID NO:68 and SEQ ID NO:69) or Figure 8 (SEQ ID NO:1-SEQ ID NO:4).

16. (Amended) The method according to claim 15, characterized in that the primers are those selected from the group of:

a) those described in Table 1 (SEQ ID NO:10-SEQ ID NO:17);

b) those described in Table 3 (SEQ ID NO:18-SEQ ID NO:67); and

c) those including the introns-exons junctions of Table 2;

d) those derived from the primers in a), b), or c).

18. (Amended) A kit for the detection of a predisposition to LGMD2 by nucleic acid amplification characterized in that it comprises primers selected from the group of:

a) those described in Table 1 (SEQ ID NO:10-SEQ ID NO:17);

- b) those described in Table 3 (SEQ ID NO:18-SEQ ID NO:67); and
- c) those including the introns-exons junctions of Table 2;
- d) those derived from the primers in a), b), or c).